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Lymphangioma: fetal neck mass, delivery of pregnancy through EXIT technique – case report, University Hospital San Ignacio (HUSI)

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Lymphangioma is a rare type of benign tumour, caused by a lack of development of the lymphatic's system components. This malformation is an accumulation of fluid mainly located in the posterolateral region of the neck in 75% of cases, followed by armpit 20%, mediastinum 1%, abdominal organs, mesenteric retroperitoneal in 2–3% & bone, lower limbs in 2%. This develops in the second and third trimester. It could be aggressive due to the infiltration in adjacent tissues or closer organs, additionally, it could be associated with chromosomal abnormalities or other malformations, basically, the prognosis depends weather it is present or not. In isolated forms, these tumours have a good prognosis. In cases where the malformation is found on the neck in the first trimester, the assessment, behaviour, name, management and etiology could be different and it is called increased NT. The prenatal management includes serial ultrasound in maternal-fetal unit, karyotyping, MRI to see the level of infiltration and the associated airway obstruction, and multidisciplinary team experienced in airways management. In cases of cervical lymphangiomas, the gestation delivery should be done by a Caesarean section (CS) with an EXIT technique in order to guarantee the fetal airway before separation of the maternal-fetal circulation. it is imperative to set up the procedure through a multidisciplinary team meeting, in order to assign each of the roles in surgery and to know the additional materials or procedures in accordance with the fetal airway compromise. In this case the diagnosis was confirmed by maternal medicine team as a cervical lymphangioma in the HUSI in Bogotá through US performed on the 34.5 week. Additionally, we used an MRI to assess the fetal airway and after words we booked meetings with an interdisciplinary team with the objective of planning the procedures and materials required during the delivery through a CS with an EXIT technique, following the institutional protocols.

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Fetoscopic urethral meatotomy in fetuses with lower urinary tract obstruction by congenital megalourethra

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Objectives: To describe the perinatal outcomes of fetoscopic urethral meatotomy in fetuses with lower urinary tract obstruction (LUTO) by congenital megalourethra.

Methods: Between 2012 and 2020, 226 cases with LUTO were referred to our fetal surgery centre in Querétaro, Mexico. We report the perinatal outcome of cases with congenital megalourethra that were selected for fetoscopic urethral meatotomy (FUM) in an attempt to release the penile urethral obstruction.

Results: Congenital megalourethra was diagnosed in 10 cases (4.4%) but only three cases with obstructive megalourethra (30%) were selected for fetal surgery. FUM was successfully performed in all three cases at a median gestational age (GA) of 21.4 (18.0–26.7) weeks and with a median surgical time of 27 (12–43) min. A resolution of urethral dilatation and subsequent reduction of the penile length and normalisation of both the bladder size and amniotic fluid were observed in all cases. The median GA at delivery was 35.1 (range 30.6–36.6) weeks. There were no fetal deaths but one neonatal death (33%) secondary to renal failure and preterm delivery.

Conclusions: Fetoscopic urethral meatotomy is feasible and is associated with good perinatal outcomes.

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Peritoneal-amniotic shunt in the management of ascites complicating bladder outlet obstruction

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Objectives: Lower urinary tract obstruction (LUTO) with anhydramnios results in pulmonary hypoplasia and neonatal demise. Renal function is assessed by fetal vesicocentesis evaluating urinary electrolytes and bladder refill in 48-72 hours, followed by vesico-amniotic shunt (VAS) placement restoring amniotic fluid and lung development. In the presence of urinary ascites, from bladder perforation, iatrogenic or spontaneous, the procedure is challenging. We sought to assess our outcomes in cases of LUTO where a peritoneal-amniotic shunt (PAS) was placed due to significant urinary ascites and anhydramnios.

Methods: Retrospective review of cases with LUTO, normal karyotype and ascites treated by PAS, followed and delivered at our centre from January 2013 to January 2020. LUTO was diagnosed by the presence of keyhole sign, hydronephrosis and anhydramnios on ultrasound.

Results: Records were available on 10 of 13 cases with one neonatal demise of a female infant from cloacal anomaly, one infant demise at 1 year age due to respiratory complications. PAS was placed either as primary procedure or following displaced VAS. Median number of shunts, PAS and VAS, was 3 (range 1-5). Average gestational age at intervention and delivery was 21.2 and 33.5 weeks respectively. Of the 8 surviving infants, 2 had end stage renal disease, 1 had a kidney transplant, 1 case was complicated by ventral hernia with incarcerated bowel at the site of shunt placement requiring surgical management. 6 of 8 surviving cases had primary PAS placement.

Conclusions: Physicians avoid PAS due to concerns for frequent displacement since the abdominal musculature is weakened by chronic distension. There is a concern for iatrogenic gastroschisis, however we only had one case of ventral hernia following VAS and not primary PAS. In addition, in our experience PAS allows for normalisation of the AFI enabling lung development and if subsequent shunting is needed, VAS may be an option if ascites has resolved with healing of bladder wall.